Movement Disorders
CLINICAL PRACTICE
RESEARCH ARTICLE

Diagnostic Performance of the "Huffing and Puffing" Sign in Functional (Psychogenic) Movement Disorders

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Abstract: We aimed at determining the diagnostic value of effort-associated behaviors ("huffing and puffing" spectrum) in patients with psychogenic movement disorders. Three blinded clinicians rated presence, severity, and duration of effort-associated features during standing and walking tasks on edited videos of 131 patients with psychogenic gait disorders and 37 with organic gait disorders. Huffing, grunting, grimacing, and breath holding were the most common effort-associated behaviors in patients with psychogenic gait disorders, with a combined prevalence of 44% and disproportionate to the severity of gait impairment, compared to organic gait disorders. The presence of huffing and puffing–type behaviors yielded a relatively low sensitivity, but high specificity, for the diagnosis of psychogenic movement disorders, increasing the odds of diagnosis 13-fold (95% confidence interval: 4.2–43.8), compared to organic gait disorders. Demonstration of effort-associated behaviors during standing and walking strongly supports the psychogenic nature of disorders when gait is involved.

The diagnosis of psychogenic (functional) movement disorders (PMD) can be challenging given the phenotypic overlap with organic disorders and the poor diagnostic agreement by clinicians using currently available diagnostic criteria.[1] Diagnostic delays result in larger accrual of disability and poor prognosis.[2] In order to formulate a positive rather than exclusionary diagnosis of PMD, signs and symptoms unique to these disorders, inconsistent or incongruent with their organic counterparts, are needed.[3]

We have observed that PMD patients with primary or associated involvement of gait and/or balance tend to exhibit verbal and physical behaviors of effort disproportionate to their disability, particularly when standing or walking. These behaviors appear to be much less prevalent among those with organic gait disorders of comparable or greater severity, such as in advanced Parkinson's disease, SCA, or motor neuron disease. We sought to examine the prevalence, phenotypic range, and diagnostic performance of such a "huffing and puffing" spectrum of behaviors in consecutively examined patients with clinically definite PMD, as compared with organic gait disorders.

Patients and Methods

The videotape records of consecutive patients diagnosed between July 2006 and August 2012 with clinically definite PMD involving gait as a primary or associated impairment, but without pain as a major symptom to avoid this source of confounding, were edited to only include segments corresponding to the standing and walking tasks documented during their neurological examination. Similar video material was collected from patients with cerebellar, spinocerebellar, and sensory ataxia during July 2011 and August 2012 (control group). One clinician rated the severity of the gait impairment combining items 27 (standing) and 28 (gait) of the motor part of the UPDRS[4] and item 16 (turning) of the Gait and Balance Scale (Appendix 1 in the Supporting Information).[5] Three clinicians blinded to subjects' diagnoses and study purpose rated these standing and walking video segments for severity, duration, and main effort-associated features: breath holding, vocalizations (moaning or groaning), grimacing, or any other manifestation of disproportionately excessive labor, herein figuratively labeled huffing and puffing (H-P). Severity was rated on a scale from 0 to 4

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(0 = none, 1 = minimal, 2 = mild, 3 = moderate, and 4 = severe). Duration was rated on a scale from 0 to 4 (0 = none, 1 = <25% of time, 2 = 25%–50% of the time, 3 = 50%–75% of the time, and 4 = >75% of the time). The total score was derived as the product of severity by duration (see Appendix 2 in the Supporting Information, see Videos 1 and 2).

Analysis Based on Combined Raters

We determined kappa agreement among three expert clinical raters for both rated cohorts. The median rank of H-P behavior across three raters was used to define the overall severity, duration, and total score. H-P was considered positive if the median score across three raters was greater than or equal to 2 ("mild"). H-P scores between PMD and control groups were compared using Wilcoxon's rank-sum test. We determined the diagnostic performance of H-P presence in classifying subjects with PMD, as compared to controls. Diagnostic performance was summarized using sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), and receiver operating characteristics ([ROC] area; defined as the average of sensitivity and specificity), including 95% confidence intervals (CIs).

Analysis Based on Individual Raters (Sensitivity Analysis)

We carried out sensitivity analysis for the H-P sign using individual raters and assessed its diagnostic performance by using five different criteria of positivity, from least to most stringent: (A) at least "mild" H-P by any one rater; (B) at least mild H-P by any two raters; (C) at least mild H-P by all three raters; (D) "moderate" H-P by any rater; and (E) "severe" H-P by any rater. $P < 0.05$ was considered significant. Data analysis was carried out using Stata 12.1 (StataCorp LP, College Station, TX).

Results

Prevalence and Phenotypic Range

One hundred thirty-one patients with PMD met criteria for inclusion. Gait was primarily involved in 10 subjects, but was associated or secondary in 121 (tremor, 50; dystonia, 21; myoclonus, 8; parkinsonism, 3; chorea, 2; mixed phenomenology, 37). Thirty-seven patients with organic gait and balance disorders were included as controls (12 SCA, 13 pure cerebellar ataxia, and 12 sensory ataxia; Table 1). Severity of gait was lower in the PMD cohort than in the organic gait cohort (3.7 ± 2.9 vs. 4.8 ± 2.7; $P = 0.04$, *t* test).

TABLE 1 Basic demographic features of the study population

	PMD (N = 131)	Controls (N = 37)
Gender (F:M)	96:35	25:12
Age at onset (years, SD)	41.5 ± 15.5	54.1 ± 17.9
Disease duration (years, SD)	9.3 ± 8.3	10.1 ± 5.1
Time to diagnosis (years, SD)[a]	4.9 ± 7.8	4.7 ± 5.1

[a]From onset of symptoms, not initial assessment at the clinic.

Huffing, grunting, grimacing, and breath holding were the most common effort-associated behaviors (Table 2). Mean total score was 4.3 (standard deviation [SD]: 5.4; range, 0–16) in the PMD group. Overall, H-P prevalence was 44% in the PMD group, whereas none in the control cohort had at least mild H-P ("minimal" H-P was scored in 25% of the control cohort). According to the five criteria for positivity, H-P prevalence in the PMD cohort was 44% (overall), 57.3% (A), 44% (B), 35% (C), 38% (D), and 17% (E; Fig. 1).

Diagnostic Performance

The kappa agreement for severity (0.38 in PMD and 0.42 in the control group) and duration (0.37 and 0.40, respectively) were relatively, low but of similar magnitude, between the groups, which allowed combination of raters for further analyses. Overall, sensitivity for the diagnosis of PMD was 44%, whereas specificity was 100% (Table 3). H-P behavior yielded a high specificity and PPV, although with a low sensitivity and NPV, given its relatively low prevalence. Compared to controls, however, H-P raised the odds of having PMD by 13 times (95% CI: 4.2–43.8).

Classifying the presence of H-P behaviors using moderate (B and C) to stringent criteria (D and E) provided high specificity, but low sensitivity. Classifying the presence of H-P behaviors

TABLE 2 H-P behaviors by frequency and association with PMD

Most common associated PMD	Huffing	Grunting	Grimacing	Breath holding	Heavy breathing	Crying, Tearing	No behavior	Total
Tremor	8/2 (5)	11/0 (1)	3/1 (0)	1/3 (0)	2/2 (0)	0/0 (0)	20	59
Dystonia	4/3 (1)	2/0 (0)	2/0 (0)	0/0 (0)	1/2 (0)	1/0 (0)	9	25
Ataxia	2/0 (1)	2/0 (0)	1/0 (0)	0/0 (0)	0/1 (0)	0/0 (0)	1	8
Parkinsonism	0/0 (0)	1/0 (0)	0/0 (0)	0/0 (0)	0/0 (0)	0/0 (0)	0	1
Myoclonus	0/0 (1)	3/0 (0)	2/0 (0)	0/0 (0)	0/0 (0)	0/1 (0)	2	9
Other[a]	1/2 (0)	2/0 (1)	1/0 (0)	1/0 (0)	0/1 (0)	0/0 (0)	6	15
Combined PMDs	7/0 (2)	2/0 (0)	2/0 (0)	1/1 (0)	1/0 (0)	1/1 (1)	5	24
Total	22/7 (10)	23/0 (2)	11/1 (0)	3/4 (0)	4/6 (0)	2/2 (1)	43	141[b]

Within each column, the first number applies to the number of subjects reported, for each of these effort-related behaviors, when *standing*; the second number applies to the number of subjects with each of these behaviors reported when *walking*. The number in parenthesis denotes the number of patients reported who displayed these effort-related behaviors when standing *and* walking, not accounted for in the first and second numbers.
[a]"Other" PMD were tics, chorea, and stuttering.
[b]There were 131 patients with a diagnosis of PMD. The sum total is 141 because 10 patients displayed a mixture of H-P behaviors.

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with less-stringent criteria (A) provided moderate sensitivity and high specificity (Table 4). Sensitivities ranged from 17% to 57%, whereas specificities range from 89% to 100%. The minimum specificity was 89% for definition A and the maximum was 100% for definitions B, C, and E. In most situations, the ROC area was >65% (range, 58%–73%). This suggested that H-P behaviors had very good discriminating ability for PMD. Across all definitions, the positive predictive performance was greater than or equal to 95% (range, 95%–100%).

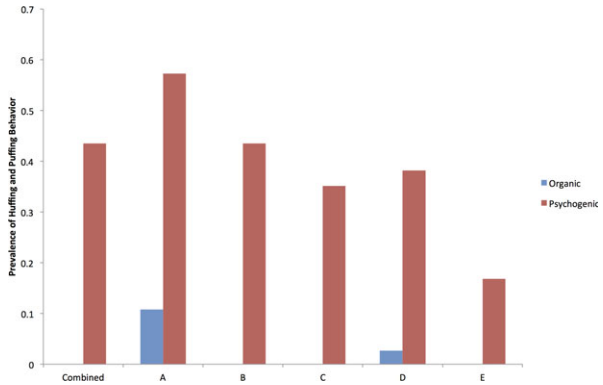

Figure 1 Prevalence of H-P in the PMD and organic cohorts. Five criteria for positivity were used, from least to most stringent: (A) at least mild H-P by any one rater; (B) at least mild H-P by any two raters; (C) at least mild H-P by all three raters; (D) moderate H-P by any rater; and (E) severe H-P by any rater. The "combined" bar represents the overall H-P prevalence without prespecified definitions. Note that mild H-P was present in controls only by definitions A and D.

TABLE 3 Diagnostic performance of H-P behavior on combined raters

Diagnostic measures	Entire PMD cohort (N = 168)	Sensitivity analysis (N = 159)
	Estimate (95% CI)	Estimate (95% CI)
Se	0.44 (0.35, 0.52)	0.57 (0.48, 0.66)
Sp	1.00 (0.91, 1.00)	0.91 (0.76, 0.98)
PPV	1.00 (0.94, 1.00)	0.96 (0.89, 0.99)
NPV	0.33 (0.25, 0.43)	0.37 (0.26, 0.48)
ROC area	0.72 (0.68, 0.76)	0.74 (0.68, 0.81)

Se, sensitivity; Sp, specificity.

Discussion

Our data suggest that PMD patients with gait involvement exhibit excessive effort-related behaviors more commonly than organic cohorts and disproportionate to the gait severity (which was actually lower in the PMD cohort). Although relatively uncommon, the presence of these H-P behaviors in patients with gait impairment substantially increases the odds of a psychogenic etiology and can thus be helpful in distinguishing PMD patients from those with organic disorders. Though the absence of this sign is common (low sensitivity), its presence is greatly supportive of the diagnosis of PMD (high specificity). Because a minimal expression of effort-related behaviors can also be present in organic gait disorders, this behavior can only be used to support the diagnosis of PMD, particularly when obvious, but does not alone serve to confirm it.

Patients with advanced neurodegenerative disorders involving gait and balance rarely "huff and puff" or "moan and groan" when standing or walking, despite a common need for a walker or wheelchair. Hence, our data give credence to the observation that demonstration of excessive effort during these tasks is *incongruent* with established behavioral patterns in organic gait disorders (at least in patients with cerebellar and sensory ataxia serving as controls in this study) and provide support toward a clinically definite category of diagnostic certainty.[6,7] The set of H-P behaviors can be considered of similar clinical value to other signs recognized as incongruent with organic counterparts, such as frequency entrainment in psychogenic tremor[8] and tonic lip deviation in psychogenic facial dystonia.[9]

Our study has a number of limitations. The "huffs, puffs, moans, and groans" are difficult to evaluate in a standardized fashion, and the threshold between "normal" and disproportionate display of effort may be in the eye of the beholder. This difficulty may have explained the relatively low inter-rater reliability, although sufficiently consistent across cohorts in the ratings of duration and severity to justify combining the data and using sensitivity analyses. In addition, the inconsistent or incongruent motor behaviors during the tasks of standing and walking likely unmasked the PMD diagnosis, and although the study hypothesis was not explicitly disclosed to the clinical raters, they may have felt more compelled to assign signs of excessive displays of effort in patients they judged as psychogenic owing to the presence of other clinical signs on video.

TABLE 4 Diagnostic performance of H-P behavior on individual raters (sensitivity analysis)

Diagnostic measures	Estimate (95% CI)				
	A	B	C	D	E
Se	0.57 (0.48, 0.66)	0.44 (0.35, 0.52)	0.35 (0.27, 0.44)	0.38 (0.3, 0.47)	0.17 (0.11, 0.24)
Sp	0.89 (0.75, 0.97)	1.00 (0.91, 1.00)	1.00 (0.91, 1.00)	0.97 (0.86, 1.00)	1.00 (0.91, 1.00)
PPV	0.95 (0.88, 0.99)	1.00 (0.94, 1.00)	1.00 (0.92, 1.00)	0.98 (0.9, 1.00)	1.00 (0.85, 1.00)
NPV	0.37 (0.27, 0.48)	0.33 (0.25, 0.43)	0.30 (0.22, 0.39)	0.31 (0.23, 0.4)	0.25 (0.19, 0.33)
ROC	0.73 (0.67, 0.80)	0.72 (0.68, 0.76)	0.68 (0.64, 0.72)	0.68 (0.63, 0.73)	0.58 (0.55, 0.62)

The following definitions were used, from least to most stringent: A: at least mild H-P by any one rater; B: at least mild H-P by any two raters (equal to median combined raters); C: at least mild H-P by all three raters; D: at least moderate H-P by any rater; and E: severe H-P by any rater.
Se, sensitivity; Sp, specificity.

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Nonetheless, it should be noted that these H-P behaviors were more common and prominent in the PMD cohort despite a lower severity of gait than the organic cohort. Stated from the other side, patients with organic gait disorders do not tend to exhibit overt manifestations of effort during standing or walking, despite advanced disability. Finally, the prevalence of H-P reported here might not be representative of unselected populations, though an effort was made to include every patient evaluated during the study period.

In summary, the presence of disproportionate H-P behaviors can be helpful to robustly support a psychogenic etiology in patients with primary or secondary gait impairment. Validation and refinement of what constitutes disproportionate manifestations of effort, prospective ascertainment of the true prevalence of such H-P phenomena in psychogenic gait disorders, but also in other psychogenic disorders (e.g., while patients attempt to perform motor tasks in the impaired body parts), and its relationship with response to therapy and other prognostic indicators are valuable targets of future research.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the First Draft, B. Review and Critique.

H.N.L.: 1B, 1C, 2C, 3A
A.K.D.: 1B, 2A, 2B, 2C, 3B
F.J.R.: 1C, 3B
A.P.D.: 1C, 3B
C.P.-J.: 1C, 3B
A.J.E.: 1A, 1B, 1C, 2C, 3B

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Supporting Information

Videos accompanying this article are available in the supporting information here.

Appendix 1. Standing and walking severity.

Appendix 2. Scale for H-P behaviors.

Video 1. H-P during (attempted) standing in a patient with psychogenic dystonia and parkinsonism with secondary gait involvement. This subject (subject 8) was rated as 4/4 ("severe") for severity and 4/4 (">75% of the time") for duration by all raters.

Video 2. Huffing during walking in a patient with psychogenic dystonia and gait involvement. Subject 25 was rated as 3.33/4 ("severe") for severity and 3.67/4 (">75% of the time") for duration.

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